



Assessment of renal involvement in patients with familial Mediterranean fever: a clinical study from Ardabil, Iran

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Key words

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Abstract

Background and Aim: Familial Mediterranean fever (FMF) is an autosomal recessive disease characterised by recurrent episodes of painful inflammation in the abdomen, chest or joints. The association between FMF and non-amyloid glomerulopathies are unusual. In this study, we describe our experiences and observations about renal involvement in patients with FMF.

Methods: A total of 108 patients with FMF was enrolled in the study. Twelve patients with FMF were referred to the Nephrology Service, for evaluation and assessment of the degree of renal involvement. All the 12 patients underwent percutaneous ultrasound-guided renal biopsies and genetic analysis.

Results: On microscopic examination of the kidney specimens, six patients were found to have amyloidosis, five focal segmental glomerulosclerosis and one patient membranoproliferative glomerulonephritis. It seems that in patients with FMF and renal amyloidosis, the response to treatment with colchicine is excellent, but in patients with FMF and focal segmental glomerulosclerosis, the response to treatment with colchicine is poor. We present an evidence-based algorithm, constructed based on literature review, to aid decision making in management of renal involvement in patients with FMF.

Conclusion: The results of our study suggest that in patients with FMF and renal involvement, non-amyloid renal lesions should be considered in the differential diagnosis in addition to amyloidosis.

Introduction

Familial Mediterranean fever (FMF) is an autosomal recessive disease characterised by recurrent episodes of painful inflammation in the abdomen, chest or joints. These episodes are often accompanied by fever and sometimes a rash.¹ Most patients with FMF experience their first attack in early childhood; in 65% of cases, the initial attack occurs before the age of 10 years, and in 90% before the age of 20 years.² FMF primarily affects populations originating in the Mediterranean region, particularly people of Armenian, Arab, Turkish and non-Ashkenazi Jewish ancestry.³ The prevalence is 1 in 256 people to 1 in 500 people among non-Ashkenazi Jews,⁴ and 1 in 1073 people among the Turkish population.⁵ The FMF gene, also named as Mediterranean Fever (MEFV) gene, is located on the short arm of chromosome 16.⁶ FMF is caused by a mutation in the gene encoding pyrin.

The exact mechanism triggering the acute attacks in FMF is unclear, but several lines of evidence point to the neutrophil as the effector of the inflammatory response at serosal surfaces.

Renal involvement is the dominant feature of FMF-related amyloidosis. It begins insidiously, causing proteinuria, then progresses to symptomatic nephrotic syndrome, and eventually ends in renal failure. The natural history of amyloidotic nephropathy in FMF is such that end-stage renal disease develops from 2 to 13 years after the onset of proteinuria.² Although the inflammatory attacks of untreated FMF are the cause of much morbidity, the major source of mortality in this disease is progressive secondary (AA) amyloidosis.⁷ Before the advent of colchicine, amyloidosis occurred in approximately 30% of Sephardic Jews and 60% of Turks with FMF.¹ Amyloidosis is much less common in Ashkenazi Jews and FMF patients living in America, including those of Armenian ancestry.⁸

The association between FMF and non-amyloid glomerulopathies is unusual. In the present study, we

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